A Comparative Cephalometric Study of the Cranial Base in Craniofacial Anomalies: Part I: Tensor Analysis

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The method of mean tensor analysis was used to study the cranial base in six craniofacial anomalies: Crouzon's disease, Apert's syndrome, Pfeiffer's syndrome, craniofacial microsomia (CFM), Treacher Collins (TC) syndrome, and frontonasal dysplasia (FND). The form was represented by five landmarks: the nasion (N), basion (Ba), sella (S), frontomaxillonasal suture (FMN), and sphenoethmoidal registration point (SE), and the deformities were computed as mean deformations from age- and sex-matched normal mean forms. The cranial base in CFM is normal in shape. The other five syndromes manifest four distinct patterns of shape variation. Only in TC and Pfeiffer's syndrome is the cranial-base angle distinctive. In Apert's and Crouzon's syndromes, point SE is displaced anteriorly upon a cranial base, small in size but otherwise normal in shape. In TC syndrome and FND, point SE is displaced posteriorly toward the sella.

The role of the cranial base in the development of craniofacial anomalies is not well understood. However, it has been speculated that cranial base abnormalities represent the primary site of the pathology in craniofacial synostosis syndromes (Moss, 1959; Kreiborg and Pruzansky, 1971; Ousterhout and Melsen, 1982). Widening of the ethmoid portion (presphenoidal portion) of the anterior cranial base has been observed in orbital hypertelorism, and abnormalities in the cranial base often accompany Treacher Collins (TC) syndrome (Rogers, 1964).

The bulk of information reported in the literature on cranial base morphology in patients with major craniofacial anomalies has been derived from measurements of dry skulls from affected individuals or from conventional cephalometric measurements on small samples of patients (Dahl et al, 1975; Herring et al, 1979; Bjork, 1972; Sperber, 1981).

In our study, the cephalometrics of the cranial base were documented in each of six groups of children with craniofacial anomalies: Crouzon's disease, Apert's syndrome, Pfeiffer's syndrome, craniofacial microsomia (CFM), TC syndrome, and frontonasal dysplasia (FND). Age- and sex-

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matched normal children from the University of Michigan reference population (Riolo et al, 1974) served as the controls. The typical pattern of difference from normal for each group studied was computed and evaluated.

Materials and Methods

Figure 1 illustrates the five cranial base landmarks used in this study. The nasion (N) is conventionally defined as the most posterior point on the frontonasal suture on the curve at the bridge of the nose. It is actually seen on the dry skull to be at an intersection of three sutures. The basion (Ba) is the most inferior posterior point on the anterior margin of the foramen magnum. The sella (S) is the center of the pituitary fossa of the sphenoid bone and is reliably found by a trained technician. Frontomaxillonasal suture (FMN) is the junction of the frontal, maxillary, and nasal bones. It is the overlapping image of a laterally symmetric pair of intersections of three sutures. Sphenoethmoidal registration point (SE) is the intersection of the sphenoidal plane with the averaged greater sphenoid wings. The location of this "point" expresses the position of two structures, not one.

The locations of the five landmarks were recorded from the lateral cephalograms of 157 individuals, all of whom had one of the six craniofacial anomalies (Table 1). From the archives at the Center for Human Growth and Development, Ann Arbor, Michigan, the same landmarks for 83 subjects from the University of Michigan University School Study were averaged in a consistent coordinate system (S–N). In the University of Michigan sample, ages ranged from 6 to 15 years. The five syndromal cases aged 5 years or less were matched to the University of Michigan means for 6 year olds; all cases aged 15 or older were matched with the University of Michigan means for 15 year olds. Because growth of the cranial base is essentially complete by age 14, little error is introduced in this step. This population provided age- and sex-specific normal mean forms of the cranial base for comparison with the study population.

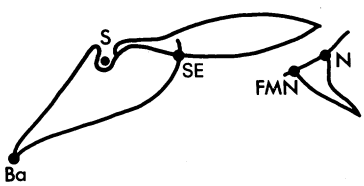

FIGURE 1 The cranial base landmarks used in this study: (N) nasion, (Ba) basion, (S) sella turcica, (FMN) frontomaxillonasal suture, (SE) sphenoethmoidal registration point.

TABLE 1. Distribution of Sample by Syndrome.

Crouzon's disease	24
Apert's syndrome	11
Pfeiffer's syndrome	4
Craniofacial microsomia (CFM)	78
Treacher Collins syndrome (TC)	9
Frontonasal dysplasia (FND)	31
	N = 157

Measurement Design: Deformation of Normal Mean into Study Population Mean

Each cranial base in the study population may be viewed as a deformation of the age- and sex-matched normal mean cranial base. The findings of the study include the distances, angles, and ratios that differ most across the comparison (Bookstein, 1983a). To facilitate the visualization of cranial base deformations in the study population, the landmarks were linked together in sets forming convenient triangles. From the five landmarks it is possible to define a total of ten triangles for analysis. For six of the ten triangles, the area proved too small for the study of deformation to be usefully employed. The four remaining triangles, which are relatively useful for analysis, are diagrammed in Figure 2.

Tensor Analysis

The most effective way to describe the deformation between a single pair of triangles (Bookstein, 1982a, 1982b, 1983) begins by considering its effect on length

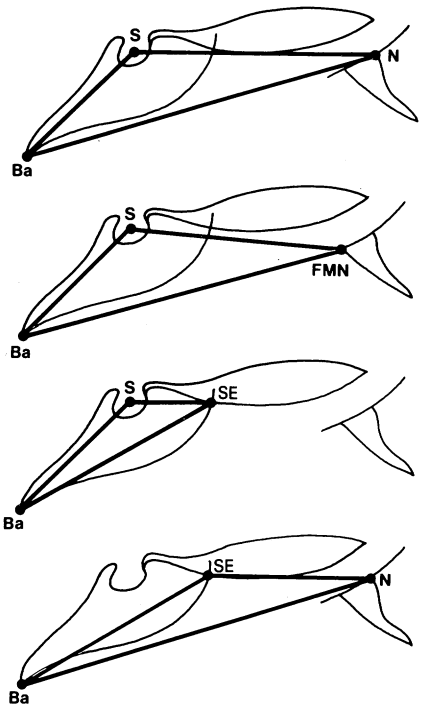

FIGURE 2 Cephalometric landmarks linked together in sets forming triangles. Deformation of the cranial base in the study population may be visualized in terms of change in distances, angles, and ratios in the landmark triangles. The quantitative comparison of these triangles was made between the normal and the study populations.

in all directions. Figure 3 depicts these effects, using lines through one point in the normal triangle ABC and through its corresponding location in a deformed triangle, its counterpart A′B′C′.

In the context of the study of deformity, the triangle on the left will be a "reference triangle", the typical or mean shape for the normative population. The triangle on the right will be a "study triangle" representing the same landmarks in a patient.

Each line drawn has a dilatation. A dilatation is a ratio of lengths: the length of the line on the right divided by corresponding length on the left, that is, deformed versus normal. Thus, dilatations are not differences (measured in mm) but quotients, with a numerator and denominator, each measured in mm. Dilatations

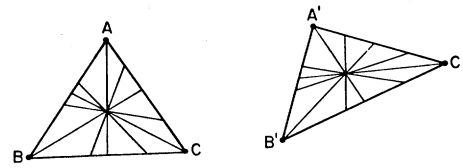

FIGURE 3 Changes in shape between two triangles may be studied in terms of length measurements made in all directions. Lines passing through a single point in the "reference" triangle ABC are compared in length to the corresponding (homologous) lines found in the "deformed" triangle A′B′C′.

greater than 1.0 represent increase of length or stretch; dilatations less than 1.0 represent reduction of length or shrink.

Circle to Ellipse: The Principal Cross

In Figure 3 the dilatations are implied as ratios of corresponding lengths between the triangles. In obtaining these ratios one may avoid the necessity for division by beginning with lines of constant length, i.e., diameters of a circle. In the deformation of a circle (Fig. 4A) one can then read the dilatation function directly.

In Figure 4, the curve on the right, which is the distortion of the circle on the left, is an ellipse. There are two facts about ellipses which tell all one needs to know about smooth shape change. An ellipse has two axes: one is the longest diameter of the form and the other is the shortest. These axes, which are at an angle of 90° to each other, are also the axes of symmetry of the ellipse. Because the diameters of the ellipse in Figure 4B represent the dilatations of the shape change depicted, these simple properties can be restated as aspects of the shape change.

Any shape change between triangles has a direction of greatest rate of change of length and a direction of least rate of change of length. These directions are at an angle of 90° both before and after transformation.

Without measuring the shapes of the triangles, one can measure the change in shape by referring to the two dilatations. They are called the *principal dilatations*; the directions along which they run are called the *principal directions* of the deformation.

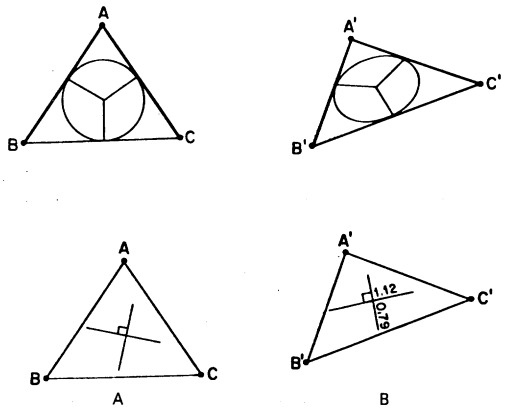

FIGURE 4 The ratios of corresponding lengths between triangles may be studied by examining the effect of the deformation on the diameter of a circle inscribed within the "reference" triangle. In this example, the circle of Figure 4A is deformed into the ellipse of Figure 4B. The ratio of lengths in A'B'C' divided by those in ABC are called dilatations (Fig. 4B). The dilatation of 1.12 indicates a stretch of 12% in the direction along the horizontal line, while the dilatation of 0.79 describes shrinkage of 21% in the vertical direction. These two axes represent the longest and shortest diameters of the ellipse found in A'B'C'. Without measuring the shapes of the triangles, one can measure the change in shape by reference to these two dilatations. They are called the principal dilatations; they align with the direction of maximum stretch and shrinkage of the form.

The representation of the two diameters of the ellipse (largest and smallest) is called the principal cross or biorthogonal cross of the shape change. In most applications, neither arm of the principal cross, which is computed to align with the shape change, will lie parallel to any side of the starting triangle. Consequently, one is recording distances not usually measured.

In this particular comparison (Fig. 4B), the direction of greatest dilatation is approximately, but not exactly, parallel to side BC of the triangle. In this direction, lengths have increased by a factor of 1.12 (a dilatation of 12%). Perpendicular to it is the direction of least rate of increase of length. In this example, the minimum is an actual decrease by a factor of 0.79, a compression by 21 percent. The area of the triangle has altered by the product of the dilatations ($1.12 \times 0.79 = 0.885$). The measure that most accurately reflects the shape change is the ratio of lengths in the two principal directions. In this case, the shape has changed by a factor of 1.12/0.79 (= 1.42).

The crosses displayed throughout this article are pictures of tensors, coordinate-free representations of geometric change. Further information about the nature of tensors is presented by Bookstein (1984).

Triangle by triangle, one can average the tensors (the crosses) for each study population in order to determine the directions of maximum and minimum average deformity from normal, i.e., the directions of greatest and least mean dilatation. By inspecting the mean tensor, one can compute the mean difference between the study population and the normals for distances or angles arbitrarily selected, as well as the net size change and net shape change. From this description of the mean differences, one can construct a set of conventional measures equivalent to them and appropriate for the clinical setting.

FINDINGS

The method of mean tensor analysis was employed 24 times, comparing the four selected triangles in the six syndromal samples to the mean "normal" form. The diagrams of principal mean dilatations and directions are illustrated in Figure 5, in which the triangles labeled A through D correspond to a composite normal mean form. Drawn over each triangle in Figure 5 are the two principal axes of deformity and the dilatations along each axis. The dilatations are printed as the fractions by which lengths in the typical syndromal case fall short of, or exceed, the matched normal means. These mean fractions together with their standard deviations are presented in Table 3 for the six syndromes and for the triangles we studied. Statistical analysis of these quantities was by the method of Bookstein (1984).

Apert's Syndrome

The four comparisons for this group (N=11) will be reviewed in detail, using the statistics associated with the principal dilatations as listed in Table 2.

The triangle Ba-S-N (Fig. 6A) shows a

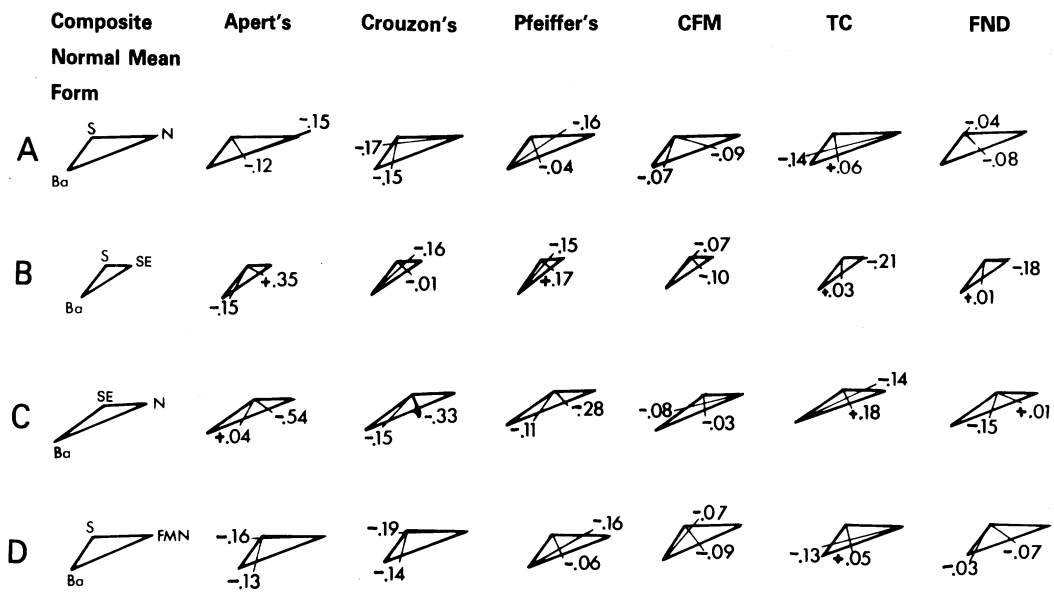

FIGURE 5 On the left are the composite mean triangles for the normal population. On the right are the principal axes of deformity and the dilatations shown along each axis. The four triangles studied (A,B,C,D) in the six syndrome populations are displayed.

compression from normal by 12 percent along the direction from the sella to a point approximately 35 percent of the distance from the basion to the nasion, and a compression from normal of 15 percent aligned nearly along the segment, basion-nasion. In this triangle, the Apert's syndrome cases show shrinkage by 12 percent to 15 percent in every direction, more a size change than a shape change. The direction of the principal dilatations (see Bookstein, 1983a) indicates that the best discrimination of Apert's syndrome from normal by a shape measure using the basion, sella, and nasion is the familiar cranial base angle. However, this discrimination is not significant for this sample of 11 patients.

TABLE 2. Statistics of Sample Variation in Apert's Syndrome* (N = 11)

Triangle	*Principal Dilatations*			
	Maximum		*Minimum*	
	Mean	*S.D.*	*Mean*	*S.D.*
Ba-S-N (A)	−.115	.159	−.147	.037
Ba-S-SE (B)	.351	.199	−.146	.101
Ba-SE-N (C)	.043	.113	−.538	.223
Ba-S-FMN (D)	−.130	.127	−.126	.038

*Additional statistics for this group are listed in Table 3.

The triangle Ba-S-SE (Fig. 6B) shows a mean relative expansion of 35 percent in one direction and a mean relative compression of 15 percent in the perpendicular direction. This combination of dilatations results in a marked change in shape, a downward and forward displacement of SE.

The triangle Ba-SE-N (Fig. 6C) is also significantly misshapen. It can be imagined that SE has been displaced forward and downward from the position it occupies in the appropriate normal population. This change is the most pathognomonic measure for the Apert's syndrome patients in this data set. The mean difference from normal is approximately seven times its standard error. As discussed earlier, SE fails to qualify as an anatomical landmark, but is an intersection of shadows generated by two separate parts of the sphenoid bone. In the anatomy of the normal cranial base (Fig. 1), the planum sphenoidale runs more or less horizontally and the ala vertically, so that the observed

TABLE 3. Tensor Statistics for the Comparison of 157 Syndromal Cranial Bases with the Center for Human Growth and Development Normals

Triangle	*Principal Dilations*				*Max−Min*	*Mean Anisotropy*	*Ratio*
	Maximum	*S.D.*	*Minimum*	*S.D.*			
Apert's Syndrome (N=11)							
Ba-SE-N	.043	.113	−.538	.223	.581	.641	.906
Ba-S-N	−.115	.159	−.147	.037	.032	.181	.177
Ba-S-FMN	−.130	.127	−.162	.038	.032	.178	.180
Ba-S-SE	.351	.199	−.146	.101	.497	.537	.925
Crouzon's Disease (N=24)							
Ba-SE-N	−.146	.056	−.330	.277	.183	.363	.506
Ba-S-N	−.146	.120	−.172	.045	.026	.223	.118
Ba-S-FMN	−.143	.107	−.186	.050	.043	.213	.199
Ba-S-SE	−.008	.180	−.156	.051	.149	.274	.543
Pfeiffer's Syndrome (N=4)							
Ba-SE-N	−.112	.045	−.281	.262	.169	.265	.637
Ba-S-N	−.035	.111	−.160	.034	.124	.169	.736
Ba-S-FMN	−.056	.120	−.158	.043	.102	.156	.654
Ba-S-SE	.166	.048	−.145	.033	.311	.316	.984
Cranofacial Microsomia (N=78)							
Ba-SE-N	−.031	.177	−.081	.047	.050	.222	.227
Ba-S-N	−.069	.099	−.086	.055	.078	.138	.128
Ba-S-FMN	−.070	.063	−.086	.105	.016	.131	.123
Ba-S-SE	−.070	.071	−.103	.169	.033	.213	.153
Treacher Collins Syndrome (N=9)							
Ba-SE-N	.179	.157	−.144	.047	.323	.362	.892
Ba-S-N	.065	.112	−.142	.030	.207	.215	.961
Ba-S-FMN	.054	.118	−.134	.034	.187	.201	.934
Ba-S-SE	.031	.151	−.215	.077	.245	.311	.790
Frontonasal Dysplasia (N=31)							
Ba-SE-N	.012	.117	−.151	.156	.163	.281	.581
Ba-S-N	−.035	.068	−.083	.111	.048	.153	.312
Ba-S-FMN	−.035	.077	−.074	.105	.039	.162	.244
Ba-S-SE	.011	.121	−.164	.101	.175	.272	.642

relative displacement of SE would be composed of a dropping of the planum in association with a forward repositioning of the ala.

When studied together, the triangles Ba-S-N and Ba-S-FMN (Fig. 6, A and D) reveal change in the relative position of FMN and N. This discrepancy is best visualized in triangle S-FMN-N (Fig. 6E). The analysis cannot determine if this represents displacement of the Apert's syndrome nasion "downward" or of the FMN "upward" with respect to normal position, as there is no other information in the vicinity.

All of these findings can be summarized in an S-N coordinate system (Fig. 7): The major cranial base anomaly observed in Apert's syndrome is the forward and downward displacement of SE. The cranial base angle (Ba–S–N), does not significantly discriminate the Apert's syndrome group from normal. In addition, the cranial base appears 12 to 15 percent shorter than normal size.

Crouzon's Disease

Analysis of the Crouzon's disease patients (N=24) demonstrates a similar shape deformation in the triangles involving SE. However, in this syndrome it was expressed more weakly. The displacement of SE toward the Ba-N line averages 33 percent of the normal separation rather than 54 percent as in the Apert's syndrome group. Again, the triangle Ba-S-N is normal in shape, but it is reduced 15 percent in size.

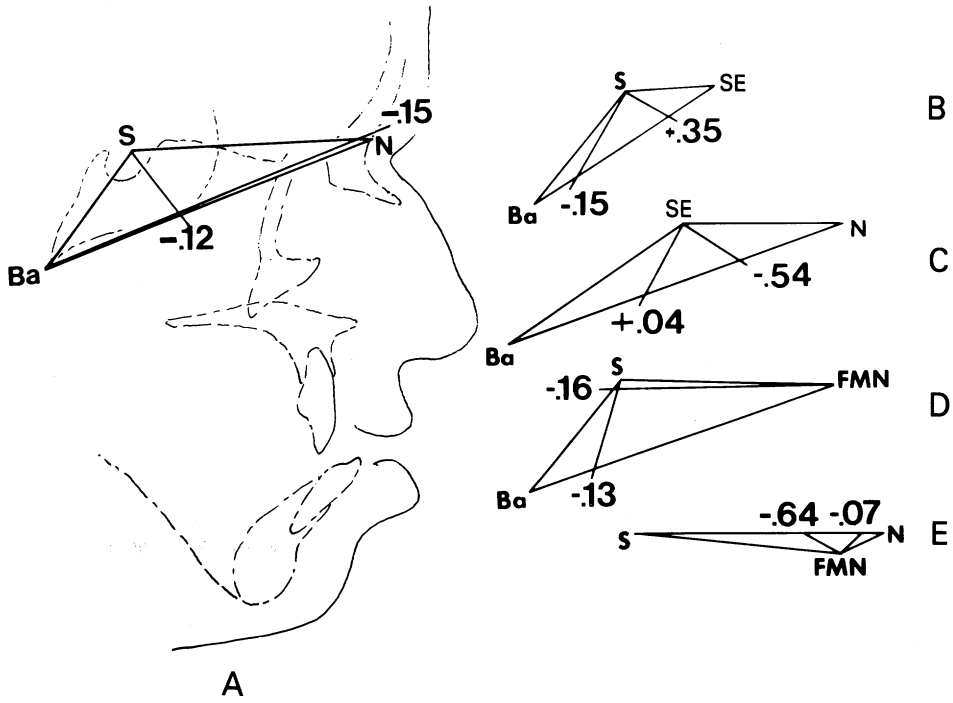

FIGURE 6 The principal mean dilatations for landmark triangles of the Apert's syndrome population.

Pfeiffer's Syndrome

In the Pfeiffer's syndrome patients, it is only the analysis of triangle Ba-S-SE that is statistically significant, owing to the small sample (N=4). The form of this triangle (Fig. 8A), is highly typical of Pfeiffer's syndrome. The triangle is deformed by a shortening of 15% ±2% along Ba-SE and an extension by 17% ±3% for the sella away from the line Ba-SE. This may be interpreted as an anteropositioning of the basion by 32 percent of its vertical distance from S-SE (Fig. 8B).

Craniofacial Microsomia

The group of 78 CFM cases shows the least shape difference from normal, although an overall reduction in size is evident. In triangle Ba-S-FMN, for example, the least abnormal direction is 7.0% ±0.7% smaller than normal, but the most abnormal is only 8.6% ±1.2% smaller than nor-

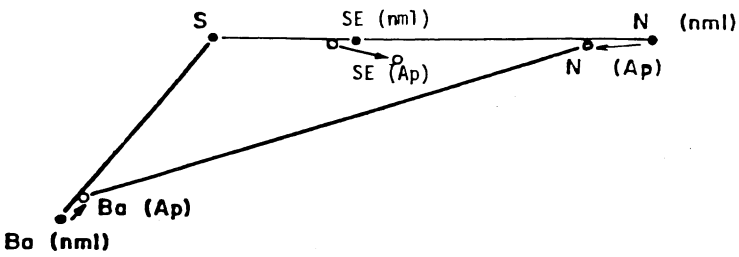

FIGURE 7 A summary of the findings in Apert's syndrome. The arrows indicate change in position of landmarks when comparing normal (nml) and Apert's syndrome (Ap) populations. SE in Apert's syndrome is displaced downward and forward. The cranial base angle Ba-S-N does not discriminate the Apert's group from normal. The cranial base triangle Ba-S-N is 12 to 15% less than normal size.

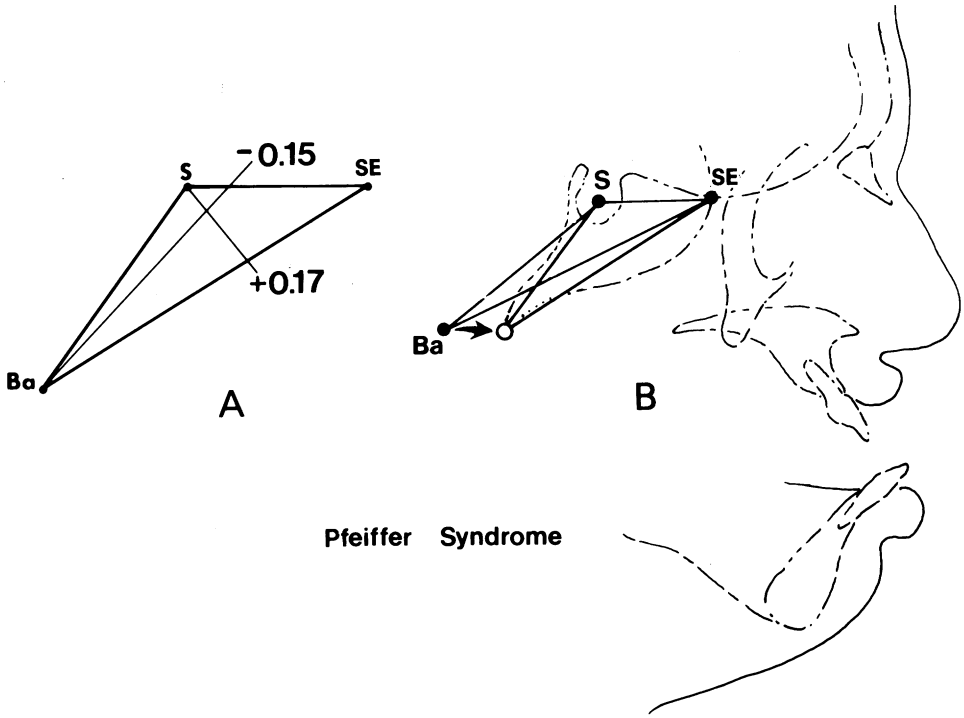

FIGURE 8 *A* The only triangle that shows statistical significance in the Pfeiffer's syndrome is Ba-S-SE. A reduction in length by 15% occurs approximately 7° off of the Ba-S axis, while the distance from S, approximately perpendicular to Ba-SE, increases by 17%.

B The deformation depicted is an anteropositioning of the basion by 32% of its vertical distance from S-SE.

mal. This triangle, although reduced in size, is not otherwise misshapen.

Treacher Collins Syndrome

By contrast, in the TC syndrome patients (N=9), all triangles are typically misshapen to a significant extent. Analysis of triangles A, C, and D, seen in Figure 5, shows the greatest shrinkage along the long axis of the cranial base, approximately from the basion to the nasion, by 14% ±1.2% with only negligible change in the perpendicular direction. Analysis of triangle B demonstrates that SE is positioned posteriorly, 21% ±3% closer than normal to the sella. The net deformation is graphically illustrated in Figure 9. As observed above in Apert's syndrome, the cranial base angle (Ba-S-N) is almost the best shape discriminator of TC syndrome. In this case, the discrimination based on this angle is highly significant.

Frontonasal Dysplasia

In the FND patients (N=31), the distortion of triangle B (Ba-S-SE) (Fig. 5) is similar to that of the TC group. SE is positioned abnormally posteriorly, 16 percent closer than normal to the sella. The triangles Ba-S-N and Ba-S-FMN, while normally shaped, were 6 percent smaller in size. The net effect is a displacement of SE more or less posteriorly in association with approximately 6 percent overall reduction in size compared to normal (Fig. 10).

Discussion

These cephalometric findings may be viewed in the light of extensive literature about the characteristic features of those craniofacial syndromes. We shall review them one by one, indicating how our results relate to the classic understanding of the underlying etiopathogenic mechanisms and their anatomical expression.

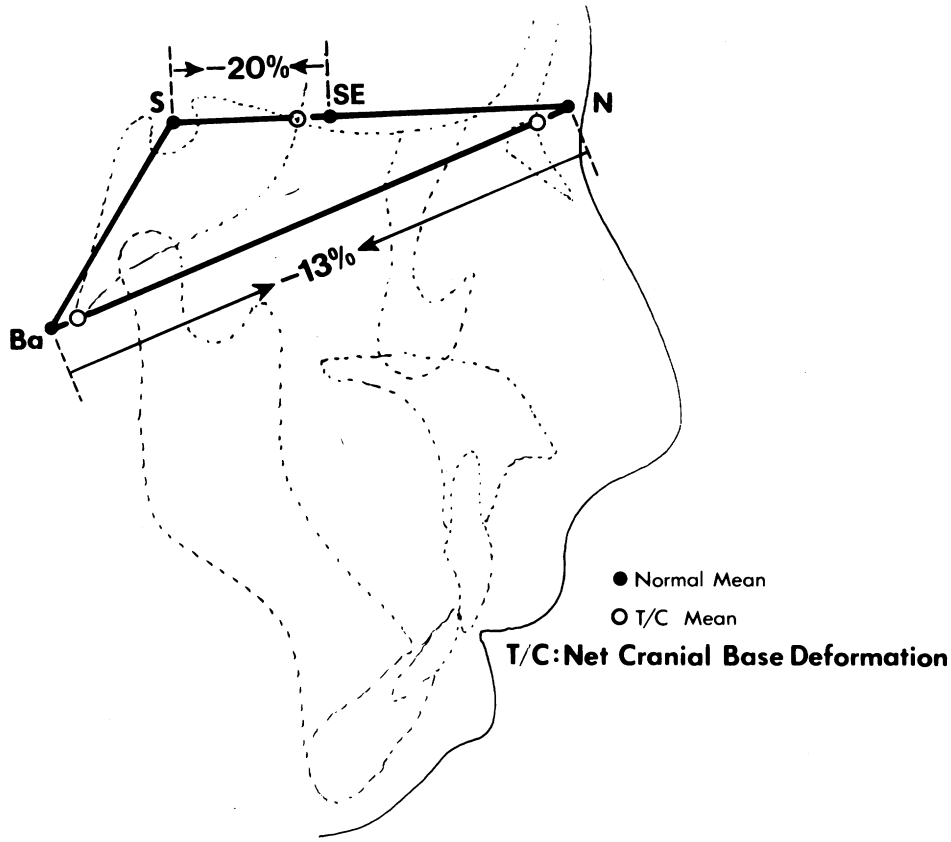

FIGURE 9 The net deformation of the cranial base in Treacher Collins syndrome. There is a shrinkage along the Ba-N axis by 13% and SE is positioned posteriorly by 20%. The cranial base angle Ba-S-N is reduced.

Craniofacial Synostosis

Numerous investigators have reported basilar kyphosis (reduced cranial base angle, Ba-S-N) in patients with Crouzon's disease (Allouche, 1935; Bornet-Ricq, 1968; Bertelli et al, 1968; Firmin et al, 1974; Castronovo, 1931), while several have noted basilar lordosis (increased cranial base angle): Vallat et al, 1958; Schmidt, 1958; Baldwin, 1968. Bertelsen (1958) found the cranial base angle to be within normal limits in the eight Crouzon's disease patients in whom it was measured. Kreiborg (1981), who studied 42 patients with Crouzon's disease, found no significant differences in the cranial base angle between Crouzon's disease patients and their age- and sex-matched normal counterparts. The same finding has been confirmed in our group of 24 patients.

The anterior cranial base (S-N) has been reported to be reduced in patients with Crouzon's disease (Castronovo, 1931; Baldwin, 1968; Kreiborg, 1981; Brenner, 1971; Ebel and Weidman, 1971; Matras et al, 1977; Horowitz, 1981; Stewart et al, 1977). Two authors have also reported shortening of the posterior cranial bases (Kreiborg, 1981; Stewart et al, 1977). The present study found a 12 to 15 percent reduction in the length of the anterior and posterior cranial bases. As in the present study, Kreiborg (1981) reported that the SE point was significantly displaced in an inferior direction. While no other investigators have reported this finding, we regard it as the main anomaly of the syndrome.

In Apert's syndrome, individual autopsy studies of affected children have reported the anterior cranial base to be nor-

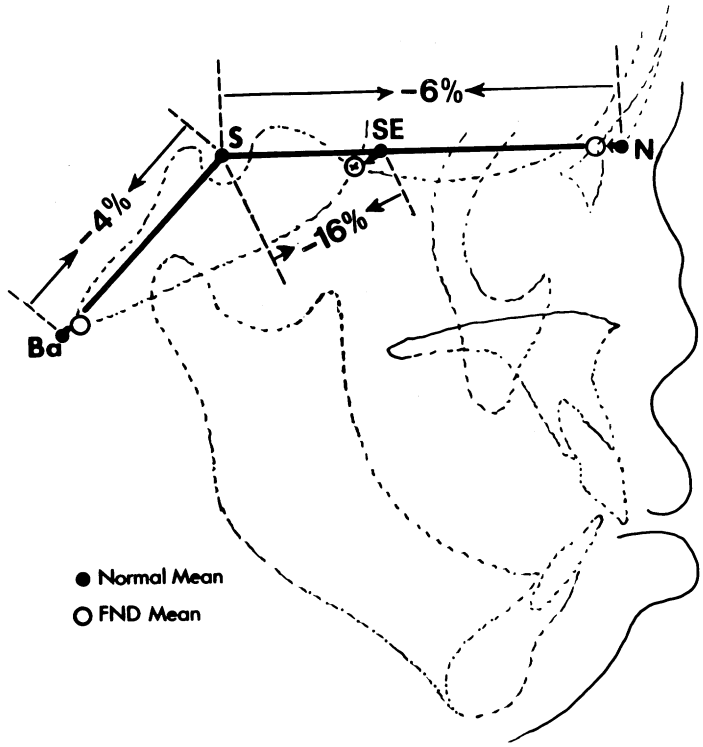

FIGURE 10 The net cranial base deformation in frontonasal dysplasia. SE is positioned posteriorly and inferiorly while the entire cranial base is reduced in size by 4 to 6%.

mal (Ousterhout and Melsen, 1982) or reduced (Kreiborg et al, 1976; Blechschmidt, 1976) in length. The cranial base angle has been reported to be greater (Blechschmidt, 1976) or smaller (Ousterhout and Melson, 1982) than normal. Kreiborg and Pruzansky (1971) reported on ten living infants with Apert's syndrome; the cranial base was reduced in all cases, especially the posterior segment. In seven of these infants with a history of strip craniectomies, the cranial base angle was normal in two, kyphotic in three, and lordotic in two. The eleven patients with Apert's syndrome in the present study demonstrated significantly shortened cranial bases with a normal cranial base angle. More importantly, the SE point was anteroinferiorly displaced.

As previously discussed, patients with Apert's syndrome demonstrated a cranial base malformation similar to that of the Crouzon's disease patients, but more pronounced. This finding has been reported by others (Kreiborg and Pruzansky, 1981). In our study, the SE point was displaced anteroinferiorly in both syndromes, but to a greater degree in Apert's syndrome. This may possibly be secondary to downward displacement of the intracranial contents from premature suture synostosis and increased intracranial pressure.

Treacher Collins Syndrome

Three necropsy reports of TC syndrome specimens (Behrents et al, 1977; Dahl et al, 1975; Herring et al, 1979) reported kyphosis of the cranial base, a finding also observed in the present study of nine patients. The decrease in the cranial base angle (kyphosis) that was demonstrated in this study is also characteristic of a number of other primary developmental defects of skeletal tissue, including cleidocranial dysostosis (Sperber, 1981). The cause has been attributed to deficient growth at the spheno-occipital synchon-

drosis. The presence of a general skeletal defect in TC patients is also suggested by the persistent intrasphenoidal synchondrosis (Dahl et al, 1975; Bjork, 1972).

Craniofacial Microsomia and Frontonasal Dysplasia

No previous cranial base cephalometric data have been reported for patients with CFM or FND. The present study showed no significant cranial base abnormality in the CFM patients, other than a diminution in size. In the FND group, the net deformation is a posterior displacement of SE in association with approximately a 6 percent reduction in overall size of the cranial base.

This cephalometric analysis of cranial base deformations not only confirms but also more accurately describes findings previously reported. The mean tensor technique, which shows principal directions and amounts of shape changes between normal and study populations, provides the investigator or clinician with a description of deformation which is independent of conventional cephalometric protocols. Instead of resembling a description of the movement of chess pieces on a chessboard, the tensor method allows one to visualize the direction and magnitude of deformation of the "chessboard" itself. This method infers the change of landmark locations as they are moved about by specifically describing deformation of the areas between them. These smooth changes in anatomy can rarely be described accurately by using routine linear and angular measurements.

Classic cephalometric analysis has not changed its complexion since the original description of the technique. One still sees attempts to describe biological contrasts and trends by arbitrary sets of distances, angles, and ratios chosen a priori. The ability to gather cephalometric data has outstripped our customary analyses of them for whatever scientific purpose. The mean tensor method provides a direct visualization of shape change, unimpeded by any predefined scheme of variables. In routine comparative morphology (contrasts, growth trends, and comparisons of growth trends within and between groups or conditions), the mean tensor technique automatically generates the distances, angles, and ratios which best characterize the effects under study.

The demonstration of tensor analysis in this paper was restricted to a limited number of landmarks in the cranial base; the authors' intention was mainly to provide a working example of the technique. Previously published findings that used conventional methods on smaller populations are, for the most part, in agreement with ours. A tensor analysis of the full face in these craniofacial syndromes is currently under way. When the tracings of patients with craniofacial anomalies are digitized and submitted to computer-assisted tensor analysis, statistically optimal and anatomically specific descriptions of the deformity become available. For example, the tensor method may be used to calculate the anatomic alterations that minimize the deformation of the resulting form from normal. In this way, the tensor method not only provides a precise vocabulary for describing shape difference but will also help in computer-aided treatment planning. If analysis of deformity does not precede the analysis of form, we will not properly appreciate how deformity originates, how it is propagated during growth, and how it is altered by clinical treatment.

SUMMARY

The six craniofacial syndromes manifest four distinct patterns of shape variation in the cranial base. (Crouzon's disease patients seem to have a mild version of Apert's syndrome in this region, while the cranial base findings in craniofacial microsomia are indistinguishable from a normally shaped cranial base of small size.) We summarize the patterns by their effects on two subsets of landmarks.

Basion, Sella, Nasion, Frontomaxillonasal Suture

In all of the syndromes except FND and CFM, the segment Ba-N or Ba-FMN is reduced approximately 12 to 15 percent be-

low normal. In Treacher Collins syndrome patients, and to a lesser extent in Pfeiffer's syndrome patients, the sella appears anomalously positioned, so that the triangle Ba-S-N is extended vertically and the cranial base angle is diminished. In the other syndromes, the cranial base triangle Ba-S-N is normal in its proportions but smaller than normal in size, by 6 to 8 percent in FND and CFM, and by 12 to 15 percent in Apert's and Crouzon's syndromes.

Sphenoethmoidal Registration Point, Sella

In Pfeiffer's syndrome patients, SE is positioned at the normal distance from the sella. In Apert's syndrome patients, SE is displaced inferiorly and especially anteriorly in relation to the sella, resulting in major disproportion. In patients with Crouzon's syndrome, the displacement is a fraction of that in Apert's syndrome patients. In patients with the other syndromes, SE is displaced more or less posteriorly toward the sella.

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